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CASE REPORT

Bilateral vocal cord abductor paralysis associated with primary herpes simplex infection: A case report

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KEYWORDS

Bilateral vocal cord
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Summary

Objective: To report a case of bilateral vocal cord abductor paralysis in the context of primary herpes simplex infection.

Case report: A 63-year-old man was urgently admitted to hospital with laryngeal dyspnoea associated with dysphagia but without dysphonia. Physical examination demonstrated the vocal cords in a paramedian position with paralysis of abduction. The patient reported primary herpes simplex infection two weeks prior to this episode. HSV serology indicated recent infection and lumbar puncture demonstrated the presence of herpes simplex virus type 1 in the cerebrospinal fluid. Complete resolution of respiratory symptoms was observed after 21 days of treatment with intravenous aciclovir.

Discussion and conclusion: Gerhardt syndrome comprises inspiratory dyspnoea without dysphonia. It used to be mainly due to syphilis, but is now mostly observed in the setting of neurodegenerative disease. The authors report a case of Gerhardt syndrome occurring after an episode of primary herpes simplex infection with the presence of herpes simplex virus in the CSF. Treatment by intravenous antiviral drugs allowed rapid resolution of the symptoms. The pathophysiology of Gerhardt syndrome remains unexplained, but the possible role of herpes simplex infection should be considered in cases of laryngeal palsy.

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Introduction

Bilateral laryngeal palsy is a serious disease. A particular form of bilateral laryngeal palsy is vocal cord abductor paralysis (VCAP) or Gerhardt syndrome, which was first described in 1863 and is characterized by severe dyspnoea

with inspiratory stridor but without dysphonia. VCAP is mostly due to neurological causes both in older and more recent reports. For example, Rebattu in 1936 reported that "all cases of vocal cord abductor paralysis should be considered to be secondary to syphilis in the absence of proof to the contrary" [1]. VCAP secondary to syphilis has become much rarer in the age of penicillin. VCAP is now essentially observed in the context of neurodegenerative disease [2,3]. We report a case of VCAP occurring after primary herpes simplex infection.

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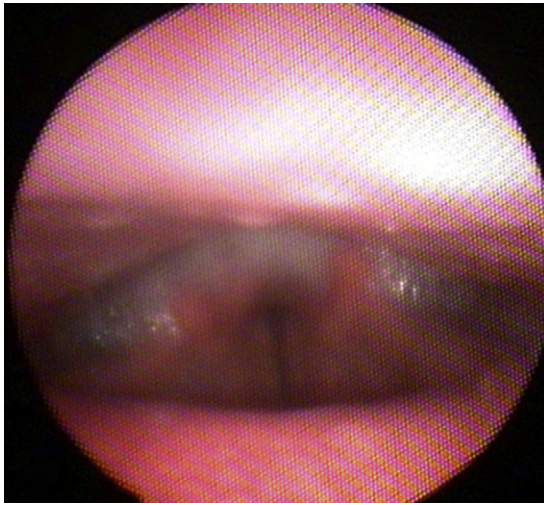


Figure 1 Initial appearance on nasal fibroscopy.

Case report

We report the case of a 63-year-old man with a history of gastrointestinal surgery, who attended the emergency room with laryngeal dyspnoea associated with dysphagia, but with no dysphonia. Two weeks previously, he had experienced an episode of febrile erythematous pharyngitis treated by oral clarithromycin (1000mg/day) for three days. Five days later, he developed gingivostomatitis with vesicular rash, considered to be primary herpes simplex infection and treated with valaciclovir 3 g daily for 10 days.

Physical examination showed the vocal cords in a paramedian position with loss of abduction but preserved adduction, associated with hypopharyngeal salivary stasis (Fig. 1). The rest of the neurological examination was normal.

The laboratory assessment was normal. Neck, chest and abdomen CT scan and head magnetic resonance imaging (MRI) did not reveal any abnormality.

The patient was admitted to hospital with symptomatic treatment by oxygen therapy and continuation of oral valaciclovir (3000 mg daily).

Herpes simplex virus (HSV1) serology showed positive IgM and positive IgG at 1500 AU/mL, in favour of recent primary infection. Varicella-Zoster virus (VZV) and cytomegalovirus (CMV) serology showed old infection. Human immunodeficiency virus (HIV), Epstein-Barr virus (EBV), hepatitis B virus (HBV), hepatitis C virus (HCV), syphilis and Lyme disease serologies were negative.

Cerebrospinal fluid (CSF) was macroscopically clear and presented laboratory signs of lymphocytic meningitis with a leukocyte count of 21/mm³, normal CSF glucose, and moderately elevated CSF protein (0.48 g/L of protein), highly suggestive of viral infection. Polymerase chain reaction (PCR) viral genomic screening of CSF was positive for HSV-1 and negative for VZV, CMV and enterovirus. The diagnosis of VCAP secondary to herpetic gingivostomatitis associated with the presence of HSV-1 in the CSF was adopted.

Antiviral therapy with intravenous aciclovir at a dose of 10 mg/kg per 8 hours was instituted for 21 days. The patient gradually improved over 10 days with improvement of

dyspnoea that persisted on effort and resolution of dysphagia allowing return home. Follow-up examination at 3 and 6 months showed normal mobility of the vocal cords with no associated symptoms and normal neurological examination, eliminating VCAP as a precursor symptom of neurodegenerative disease.

Discussion

VCAP or Gerhardt syndrome has also been previously described as "respiratory paralysis of the larynx" [1]. The characteristic clinical features consist of severe dyspnoea exclusively on inspiration, associated with little or no voice changes. Laryngeal examination reveals vocal cords with a normal morphology in a closed or paramedian position. VCAP is usually observed in the context of neurodegenerative disease. Apart from the classical "syphilitic tabes" [1], cases of VCAP have been reported much more recently in Parkinson's disease, progressive supranuclear palsy, amyotrophic lateral sclerosis, autosomal dominant cerebellar ataxias and chronic inflammatory demyelinating polyneuropathy [2–5]. Few cases of VCAP secondary to infection have been reported apart from those related to influenza virus [6], and HSV [7].

HSV is a DNA virus and two types of HSV have been described: HSV-1 and HSV-2. Viral transmission is interhuman by direct mucocutaneous contact (saliva and skin lesions). Primary HSV infection represents a period of intense viral replication. The virus then spreads along nerve trunks by retrograde axonal transport to cranial or spinal sensory ganglia [8]. Primary infection is followed by a phase of latent infection, associated with no clinical features and no viral replication. Reactivation of HSV infection occurs when the virus travels along the nerve to the skin or certain organs, especially the central nervous system, where it can cause meningoencephalitis [8]. Herpes encephalitis comprises an infectious syndrome, a meningeal syndrome, and an encephalitic syndrome (disorders of consciousness, seizures, neurological defects (monoplegia or hemiplegia, cranial nerve palsy, myelitis or abnormal movements), and behavioural and memory disorders).

Lumbar puncture was performed in the case reported here due to the probable focal neurological lesion (tenth cranial nerves) occurring after an episode of herpetic gingivostomatitis. Due to the presence of HSV in the CSF, a diagnosis of herpetic VCAP with laboratory signs of HSV meningitis was proposed. The patient was therefore treated with intravenous aciclovir, as recommended for herpes encephalitis [8].

Cases of herpetic laryngeal palsy have been rarely reported in the literature [7,9,10]. An interesting case was that of a 52-year-old man with diabetes and a history of smoking [7]. This patient presented a similar clinical history to that observed in the present case, comprising laryngeal dyspnoea associated with dysphonia and paroxysmal cough, following an episode of herpetic gingivostomatitis. Physical examination revealed VCAP with inflammation of the vocal cords. The morphological abnormalities of the vocal cords resolved with 24 hours in response to corticosteroids, antihistamines and antibiotics, while laryngeal palsy persisted. The patient presented positive HSV serology. Imaging (CT

and MRI) was normal. An improvement was observed after 4 weeks of antiviral therapy (oral aciclovir 1000 mg/day) with complete resolution of symptoms at 6 months. Lumbar puncture was not performed. Electromyography revealed fibrillation potentials at rest, indicating a peripheral nerve lesion. Laryngeal electromyography would have been useful in our case in order to define the level of the neurological lesion. One way of confirming the herpetic origin was proposed by Flowers in a case of vagal mononeuritis with right vocal cord paralysis, suspected to be due to herpes simplex virus [10]. Biopsy of the affected vocal cord demonstrated the presence of HSV. Biopsy was not performed in the present case due to the normal appearance of the vocal cords.

A diagnosis of Gerhardt syndrome accompanied by HSV meningitis following primary herpes simplex virus infection was therefore adopted. The pathophysiology nevertheless remains uncertain, as the presence of HSV in the CSF and the normal appearance of the vocal cords (no signs of laryngitis in contrast with Pou and Carrau [7] and Flowers and Kernodle [10]) were in favour of a central nervous system lesion, while the normal neurological examination and normal brain imaging excluded a diagnosis of herpes encephalitis. There were no arguments in favour of a peripheral nerve lesion. In conclusion, several arguments indicate a link between VCAP and herpes simplex virus infection: clinical (onset of VCAP soon after an episode of herpetic gingivostomatitis), laboratory (HSV serology indicative of primary herpes simplex infection and positive HSV PCR in the CSF) and therapeutic (rapid improvement over 10 days).

Conclusion

We report a case of VCAP probably associated with herpes simplex virus infection. Although the causal relationship was not formally demonstrated, clinical and laboratory arguments and the marked improvement in response to antiviral therapy are in favour of the herpetic origin of VCAP. VCAP is a complex disease in terms of its aetiology and pathophysiology, which has not been fully elucidated. This case illustrates the value of lumbar puncture in some cases of

vocal cord paralysis, but also raises the question of the use of antiviral agents in cases of laryngeal paralysis considered to be idiopathic.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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